Klippel and Trénaunay's Syndrome

768 Operated Cases

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Since 1945, we have operated on 786 patients with Klippel and Trénaunay's syndrome. Elongation of the impaired limb was invariably found while edema was present in 84%, varicose veins in 36%, and flat angiomata in 32%. Venography and surgical exploration have demonstrated malformation of the deep veins involving the popliteal vein in 51%; superficial femoral vein, 16%; both popliteal and superficial femoral veins; 29%; iliac veins, three per cent; and lower vena cava, one per cent. Good clinical results have been achieved following the surgical release of these deep veins in the lower limb. During childhood, when the difference in limb length is noteworthy, ligature of the popliteal vein of the shorter limb induces a compensating elongation. Klippel and Trénaunay's syndrome may be associated with lymphatic malformations, including lymphedema and malformation of the lymph vessels. Knowledge of the pathophysiology of these malformations of the deep veins enables a better understanding of the clinical manifestations of the condition, as well as the improved treatment of the serious vesical or rectal hemorrhage which occurs in one per cent of these patients.

IN 1900, KLIPPEL AND TRÉNAUNAY[1] described a syndrome of osteohypertrophic varicose nevus, which now bears their name. This syndrome is characterized by: (1) a more or less extensive nevus on the involved limb, (2) varicose veins on the involved limb arising in childhood, and (3) hypertrophy of all of the tissue and especially of the bones in the impaired limb.

Klippel and Trénaunay emphasized that the nevus is almost invariably present and that it is the fundamental problem that leads to other complications. They provided an excellent historical review of the nevus and described a form of the disease without a nevus. In addition, these authors described a third type that they called crossed and dissociated, and in which the hypertrophy of the varix is present on one limb but the vasular nevus is located on the opposite limb. Following this initial work, many additional observations were reported: Apert, 1909; Leroux and Raoul Labbé, 1910; Lance, 1921; Babonneix and Lance, 1924; Roederer and Phillipon, 1927; and Apert, 1931.

In 1918, Parkes Weber[2] described "hemangiectatic hypertrophy" in which all hypertrophies of the lower limb were included, associated not only with varix as in Klippel and Trénaunay's syndrome, but also those associated with congenital arteriovenous fistulas. This report confused some observers who only considered complications resulting from congenital arteriovenous fistulas when, in fact, many of these problems resulted from Klippel and Trénaunay's syndrome.

We were unaware of Klippel and Trénaunay's syndrome when in 1943 we examined by means of venography a typical patient, who had been presented by Professor Apert at the "Societé Française de Pédiatrie" in 1931. The patient was a 16-year-old female and had been referred for edema of the right leg (circumference of the calf, +9 cm; circumference of the thigh, +8 cm; elongation of the right lower limb, +5 cm) and the presence of a vascular nevus on the lateral aspect of the thigh and numerous varices. Venography (Fig. 1) revealed the absence of contrast medium in the popliteal vein as well as two large collateral veins on each side of the knee. Arteriography was normal. At the time of operation on December 26, 1943, the mid-portion of the popliteal vein was compressed by a fibrous band (Fig. 2), and division of this band resulted in dilation of the vein. Fourteen years later, the edema was not very evident, the difference in the length of the limbs was reduced from 5 to 3.6 cm, and the diameter of the popliteal vein, which had been 1–1.5 mm at the time of operation, had increased to 5 mm in diameter. Thus, in this patient it was possible to demonstrate, for the first time, compression of the popliteal vein in Klippel and Trénaunay's syndrome. Surgical division of this band compressing the deep veins improved the clinical symptoms

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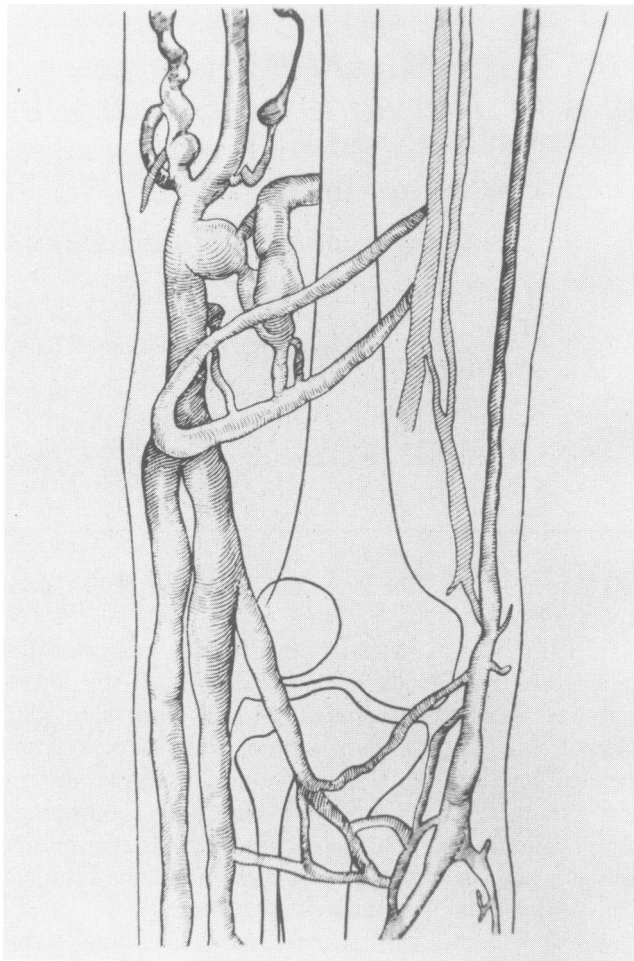

FIG. 1. Venography revealed the absence of contrast medium in the popliteal vein as well as two large collateral veins on each side of the knee.

considerably. These initial findings, as well as their value, were confirmed in an additional 767 patients whom we observed and operated on during the next 40 years.

Methods and Results

From December 1943 until July 1976, we operated on 614 patients with the Klippel and Trénaunay's syndrome. The location of the venous abnormalities discovered during these operations is presented in Table 1.

From July 1976 until July 1983, 154 other patients with malformations of the deep veins underwent surgical procedures. Thus, we studied and operated on a total of 768 patients with Klippel and Trénaunay's syndrome. During the same period, 32 patients with congenital arteriovenous fistulas of the limbs and 83 patients in whom phlebitis developed in childhood were operated upon. The two latter groups were characterized by an edematous and elongated limb, as well as venous hypertension of the involved limb.

Our examination technique for Klippel and Trénaunay's syndrome has remained unchanged. A thorough clinical examination demonstrates the extent of the edema, the edge of elongation of the limb, the existence of flat angiomata, the presence of varicose veins on the limb or abdominal wall, and finally the absence of a thrill or continuous bruit. The foot is often longer and thickened. Vaginal, rectal, or vesical hemorrhages may have occurred, as well as bleeding at the surface of the angioma, lymphorrhea, or an attack of lymphangitis. The clinical examination is followed by x-rays of the bones of each limb. Skeletal length is recorded as well as measurements of the circumference of the thigh, calf, ankle, and foot of each leg at the same level.

Venography is performed and is repeated during the postoperative follow-up. The venous blood pressure is determined by an electromanometer which usually shows a flat curve, in distinction to an arteriovenous fistula that produces a dampened arterial or systolic type of curve. The induced hyperlipidemia test[4] enables recognition of an associated malformation of chyliferous vessels. Finally, the operative report includes a diagram of the lesions found.

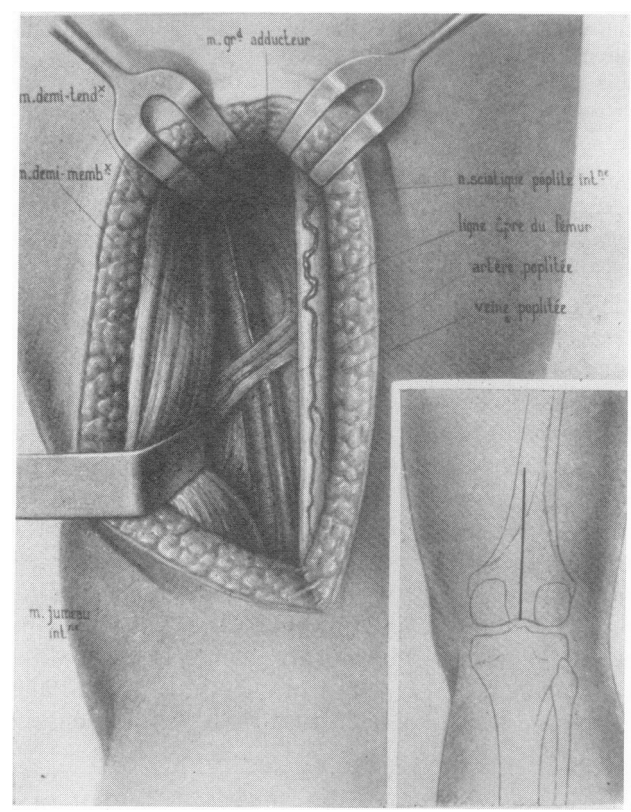

FIG. 2. The mid-portion of the popliteal vein was compressed by a fibrous band, and division of this band resulted in dilation of the vein.

Symptomatology of Klippel and Trénaunay's Syndrome

The symptomatology as described by Klippel and Trénaunay is remarkably precise. The osteohypertrophic varicose nevus, which is the most complete type and easy to recognize clinically, occurs in only 32% of our patients. The frequency of the different symptoms in our first 614 patients is depicted in Table 2.

In 1945, we noted the invariable presence of osseous elongation in Klippel and Trénaunay's syndrome. At that time, we also noted the elongation of the involved limb in two patients with sequelae of phlebitis beginning in childhood (0.5–2 cm) and in three patients with congenital arteriovenous fistulas in an extremity (1–2 cm). Venous hypertension exists in three of these disorders resulting from (1) a blockage of the deep venous channels in Klippel and Trénaunay's syndrome and in the sequelae of childhood phlebitis and (2) an abnormal inflow of arterial blood in the veins of patients with congenital arteriovenous fistulas.

Following our first clinical observations, we experimentally verified the effect of venous hypertension on osseous growth. In seven dogs aged 1 month, the veins of the right hindleg were ligated at the groin or in the popliteal region. After 12 to 18 months, the elongation of the leg varied from 2.6 to 7.6%.[5]

In the child or adult, the presence of a difference in length between two limbs suggests the need for venography of the longer side preparatory to an operative procedure if a venous malformation is found. We have operated upon 48 children between 6 and 12 years of age with a pronounced difference in the length of the two limbs. Each underwent venous freeing of the longer side and ligation of the popliteal vein of the shorter side. The difference in length between the two limbs was greatly reduced or even absent by adult life.

Edema of the impaired limb, which was observed in 84% of our patients, may be present at birth, appear at puberty, or even appear in adult life. It is often more apparent below the knee. When the thigh is large and edematous, there may be an association between the Klippel and Trénaunay's syndrome and common lymphedema or reflux of chyle in the lymphatics of the leg. In general, the edema is largely reduced after freeing of the venous malformation. However, this is not an absolute rule and these patients should be followed annually, especially when operation is performed at a very early age. Two case reports involving patients with uncommon but illustrative conditions we presented.

Case 1. At the time of operation on an 8-year-old girl, agenesis of the popliteal vein was found and the popliteal artery was covered by a lymphatic sheath. Fifteen years later, this patient suffered from true elephantiasis of the leg extending barely above the knee. Complete superficial lymphangiectomy below the knee produced a good result, which has been maintained for 10 years.

TABLE 1. *Localization of the Venous Abnormality in 614 Operated Klippel and Trénaunay Syndromes*

Lower limb	489
Upper limb	80
Lower and upper limbs	20
Four limbs	13
Both lower limbs	12
Total	614

Case 2. A 3-year-old girl was operated upon for compression of the popliteal vein by a band. Without venography or lymphography, she underwent 18 successive operations elsewhere for elephantiasis of the two lower limbs. A large amount of chyle drained from the unhealed incisions. Lymphangiography revealed huge, chyle-filled lymphatics. The induced hyperlipidemia test confirmed a malformation of the chyliferous vessels. Thus, in this case, Klippel and Trénaunay's syndrome was complicated by a bilateral lymphedema with reflux of chyle in the lymphatics of the leg.

Varicose veins develop and serve as substitute channels for the obstructed deep veins and were clinically manifest in 36% of our patients. Although venography showed that they were almost invariably present, they are often concealed by edema. The suprapubic varicose veins should never be resected as they serve as substitute channels for the atretic iliac veins. A varix located on the lower limb is resected only after relieving obstruction of the deep veins. When varicose veins are idiopathic and have been present from birth, it is necessary to determine whether one of the lower limbs is longer than the other. Venography should always be performed in the patient with unexplained elongation of the lower extremity.

Flat angiomata were present in only 32% of our series of Klippel and Trénaunay's syndrome. They often appear as purplish-red or dark purple areas with a verrucous texture that may bleed upon contact. Large varicose veins may underlie the skin. Some lesions are light pink without a change in skin texture. This cutaneous spot is very important in the diagnosis; however, it is absent in 68% of cases.

The trophic changes observed in ten per cent of our patients of Klippel and Trénaunay's syndrome are secondary to the venous stasis. These changes improve with operative relief of the venous obstruction.

Characteristics and Site of Obstruction in the Klippel and Trénaunay's Syndrome

Of the 614 operated cases, 559 procedures were performed on the lower limb. In Table 3, the localization

TABLE 2. *Frequency of the Different Symptoms in Klippel and Trénaunay's Syndrome (614 Cases)*

Elongation	Invariably present
Edema	84%
Varix	36%
Flat angiomata	32%
Trophic changes	10%

TABLE 3. *Klippel and Trénaunay Syndrome (559 Operations on the Lower Limbs)*

Only popliteal vein	284 (51%)
Only superficial femoral vein	88 (16%)
Popliteal vein plus femoral vein	164 (29%)
Iliac veins	19 (3.3%)
Lower vena cava	4 (0.7%)
Total	559

of the obstruction in these patients is presented. Lesions of the popliteal vein (284 isolated lesions, plus 164 combined popliteal femoralo malformations, a total of 448 operations) are shown in Table 4.

Malformations of the superficial femoral vein were verified 252 times (88 isolated and 164 associated with compression of the popliteal vein). These observations are summarized in Table 5. Atresia is more often observed in the superficial femoral vein (40.8%) where it often affects the entire length of the vein rather than in the popliteal vein alone (7.3%).

There are two types of malformation of the iliac veins as shown in Table 6. Malformations of the lower vena cava are represented in our series by four cases of agenesis of the lower vena cava below the renal veins. The latter usually join to form the upper part of the vena cava. The lesions in the 80 patients operated upon for malformation of the brachial and axillary veins are summarized in Table 7.

Pathophysiology

Many pre- and postoperative venograms combined with operative findings have enabled us to determine the pathophysiology of Klippel and Trénaunay's syndrome. In the malformations of the popliteal vein, the venous drainage is maintained around the knee by the development of collaterals. These include a large great saphenous vein medially, large venous channels leading to the outer surface of the articulation laterally, and lastly, in the midline by development of a large vein of the sciatic nerve which drains part of the deep veins of the calf. The latter vein usually drains into the internal iliac vein often passing into the pelvis *via* the sciatic and gluteal notches.

In the case of atresia or very tight compression of the superficial femoral vein, the substitute circulation is provided by a very large greater saphenous vein, by a large vein of the sciatic nerve, by a large retro-adductor vein which drains from above into the large deep femoral vein, and lastly, by large subcutaneous veins that develop on the lateral surface of the thigh. The vein of the sciatic nerve joins the large varicose veins from the lateral surface on the thigh behind the greater trochanter forming a large venous trunk which then divides into two branches. These pass into the pelvis—one through the sciatic notch, the other through the gluteal notch—and finally both branches connect to the internal iliac vein which becomes very voluminous due to this large inflow of venous blood.

TABLE 4. *Lesions of the Popliteal Vein (448 Operations)*

Agenesis	30 (6.6%)
Hypoplasia	33 (7.3%)
Perivenous sheath	66 (14.7%)
Compressions and fenestrations	319 (71.2%)

TABLE 5. *Lesions of the Superficial Femoral Vein (252 Operations)*

Agenesis	7 (2.7%)
Hypoplasia	103 (40.8%)
Sheath	5 (1.9%)
Compression and fenestration	137 (54.3%)
Total	252

During operation in patients with atresia or tight compression of the superficial femoral vein, we typically find a large common femoral vein above and a small superficial femoral vein below. This vein must be freed along its entire length. The large deep femoral vein must also be freed for its entire length by completely retracting the adductor magnus muscle flush with the femur. In this way, the large retro-adductor vein is also progressively freed. This vein has several large anastomoses to the vein of the sciatic nerve which should also be freed.

Before operation, the substitute venous circulation is formed by an anterior system consisting of the great saphenous vein and by a dilated deep femoral vein, both terminating in the common femoral vein. The posterior system is comprised of the vein of the sciatic nerve and by the dilated veins which develop on the lateral surface of the thigh. These drain into an enormous internal iliac vein (14–22 mm in diameter). Due to this circulatory overload, the collateral veins of this very dilated internal iliac system (vesical, genital, and rectal veins) are not able to drain normally. Dilated varicose veins appear on the bladder, internal genital organs, and the rectum. These large varicose veins may rupture into the bladder (hematuria), into the vagina, or into the rectum (repeated rectal hemorrhages in a very septic environment). Eight of our patients suffered from these hemorrhagic complications and one died from repeated rectal hemorrhage complicated by septicemia, while two others underwent partial cystectomy with good results, and two others underwent resection of the rectum with excellent results.

TABLE 6. *Lesions of the Iliac Veins (19 Operations)*

Agenesis	7 (36.8%)
Hypoplasia	12 (63.1%)
Total	19

When confronted with these vesical or rectal hemorrhages the first step in therapy is to free the superficial femoral vein and especially the deep femoral vein and its connections with the retro-adductor vein. The latter two veins form an anastomosis between the anterior and posterior venous systems. Upon freeing these two veins, a large portion of the blood from the posterior system empties into the anterior system, thus reducing the circulatory overload in the dilated internal iliac vein. Three of our eight patients suffering from vesical or rectal hemorrhages were cured by this vascular technique.

The malformations of the iliac veins may be isolated or associated with malformations of the femoral or popliteal veins. The substitute channels are similar to those in iliac phlebitis, especially with a dilated suprapubic vein diverting the blood from the impaired limb toward the common femoral vein on the opposite side. The venous blood from the malformed side also flows toward the healthy side through the outer genital veins, as well as toward the thorax and the upper vena cava through the veins in the abdominal wall. Often the veins in the outer surface of the hip return through the gluteal and obturator veins, either toward the internal iliac or the middle sarcal vein, or directly toward the lower vena cava.

Klippel and Trénaunay's Syndrome Associated with Lymphatic Malformations

Three groups of lymphatic malformations may be associated with Klippel and Trénaunay's syndrome.

1. *Lymphatic malformations resulting from the obstruction of the deep veins.* When a fibrous band compresses a femoral or popliteal vein, it also disturbs the deep lymphatic vessels that accompany this vein. During operation, a group of large lymphatic varices are found below the compression. After division of the obstructive lesion, the vein expands and the lymphatic varices collapse.
2. *Klippel and Trénaunay's syndrome associated with a common lymphedema.* This association has three different clinical aspects:
   (A) A child suffers from minor edema of the lower limb and this leg is longer than the other. Venography reveals the deep venous malformation which is surgically corrected. The edema is reduced for several years and then again increases. As an adult, there is a true lymphedema that needs to be operated on after verification by venography that the deep venous circulation has returned to normal.
   (B) In the second group, a child may present with a much more severe edema, especially in the thigh, than is typically found in Klippel and Trénaunay's syndrome. However, this limb is longer. After venography the venous malformation and the lymphedema should be operated upon. In 1954, we operated on a 12-year-old girl suffering from Klippel and Trénaunay's syndrome with extensive edema. The sciatic nerve was found to be considerably enlarged and a biopsy of the perineural tissue resulted in a diagnosis of Dejerine-Sottas' syndrome. In 1959, we operated on the lymphedema using our technique. Twenty-three years later, the limb is normal and the Dejerine-Sottas' syndrome has not recurred.
   (C) In the third group, the sympatomatology indicates a lymphedema. However, upon examination a distinct elongation of the edematous limb is discovered. An example of this was a child who suffered from lymphangitis every 3 or 4 months beginning at the age of 2. From 6 years of age on, severe lymphorrhea appeared weekly. We examined him when he was 21 years old. He had

TABLE 7. *Malformations of the Brachial or Axillary Veins (80 Cases)*

Agenesis	12 (15%)
Hypoplasia	18 (22.5%)
Compression	50 (62.5%)
Total	80

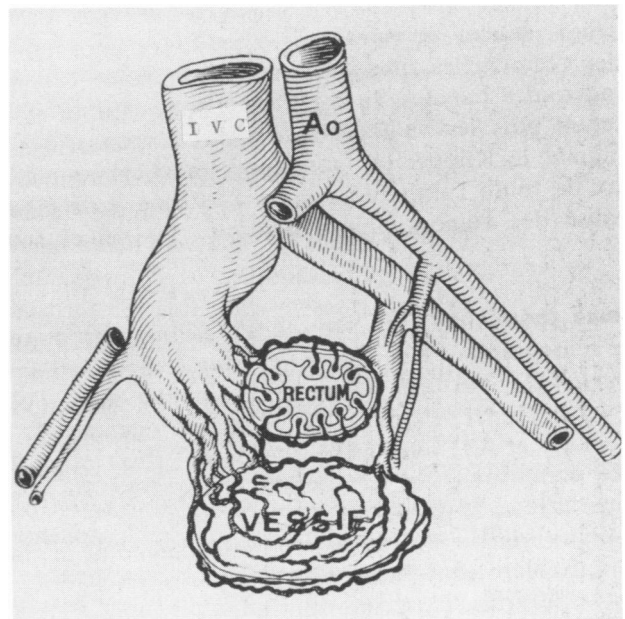

FIG. 3. Diagram explaining the physiopathology of rectal or vesical hemorrhages.

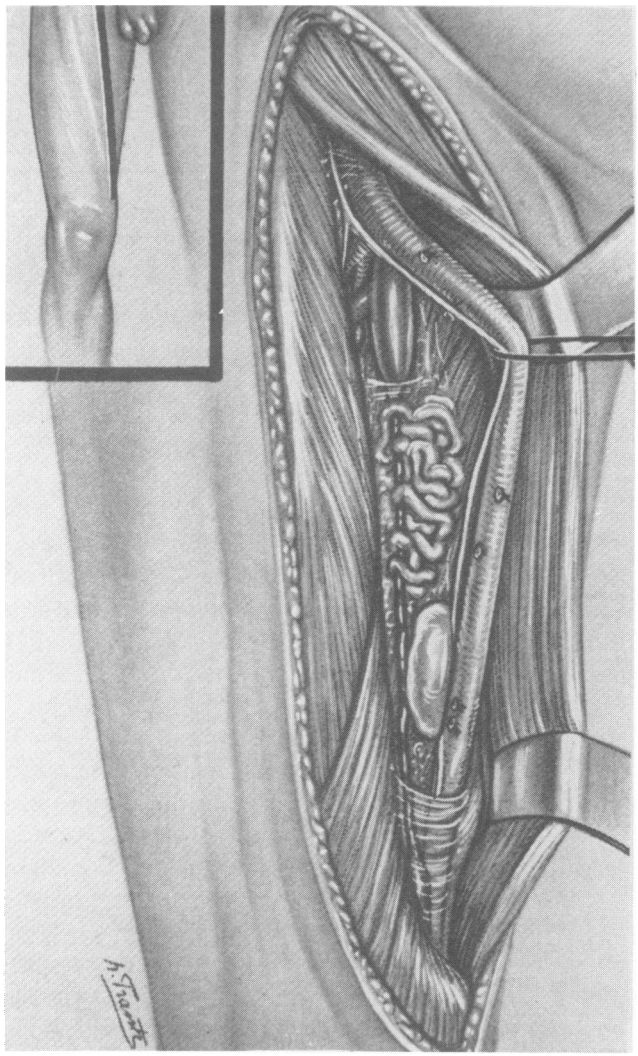

FIG. 4. Compression of the superficial femoral vein and of the femoral lymphatics. Large lymphatic varix.

extenisve edema of the limb with an elongation of 7.5 cm. Venography revealed a malformation of the superficial femoral vein. During operation (Fig. 5), we found agenesis of the superficial femoral vein and a dilated retro-adductor vein (Fig. 6). Lymphography revealed extensive dilatation of the lymphatics of the whole lower limb (Fig. 7) and of the iliac region. This observation is pointed out in our color film on Klippel and Trénaunay's syndrome.

3. *Association of Klippel and Trénaunay's syndrome with malformation of the chyliferous vessels.* In November 1945, we laid bare the small saphenous vein behind the peroneal malleolus in order to perform venography in a 33-year-old woman suffering from an extensive edema of the lower limb. Upon exposure of this vein, we found a large white lymphatic channel and 55 ml of chylous fluid, which was removed and replaced with an opaque medium. Radiography revealed a very large dilatation of the deep and superficial lymphatics of the leg, as well as reflux of opaque medium into the lymphatics of the foot. In 1943, we performed lymphangiography of the superficial lymphatics, and for the first time obtained radiographs of the deep lymphatics and established the diagnosis of lymphedema with reflux of chyle in lymphatics of the leg.

In June 1948, the same technique of exposing the same saphenous vein with discovery of a large, white lymphatic enabled venography, as well as lymphangiography in a 13-year-old boy with a leg 6 cm longer than the other. The venogram revealed a malformation

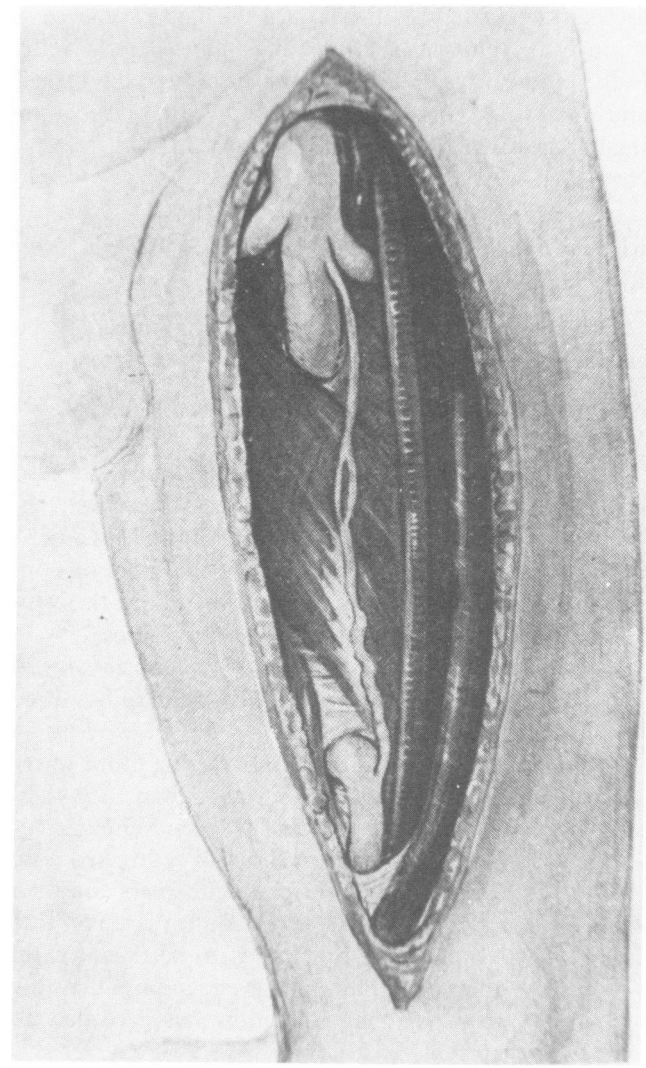

FIG. 5. Agenesis of the superficial femoral vein.

of the superficial femoral vein, and lymphangiography showed very large lymphatics in the lower third of the leg with reflux of the opaque medium into the lymphatics of the foot. We first divided an obstructing band that compressed the superficial femoral vein. One year later, our technique of total superficial lymphangiectomy followed by lateral resection of the dilated chyle-filled iliac lymphatics yielded a good result which has been maintained for 24 years. Here, Klippel and Trénaunay's syndrome is associated with lymphedema and reflux of chyle in the lymphatics of the leg, which are responsible for the lymphorrhea.

In 1951, after venography, we divided an obstructing band on the popliteal vein in an 8-year-old boy with edema of the lower limb and with a 2.5 cm limb elongation, a typical Klippel and Trénaunay's syndrome. Six months later, a left chylothorax became evident. Thoracotomy was performed and a ruptured lymphatic sutured. In 1956, a chyluria became evident and recurred during a 3-year period. In 1957, we documented edema of the genital organs with white cutaneous vesicles

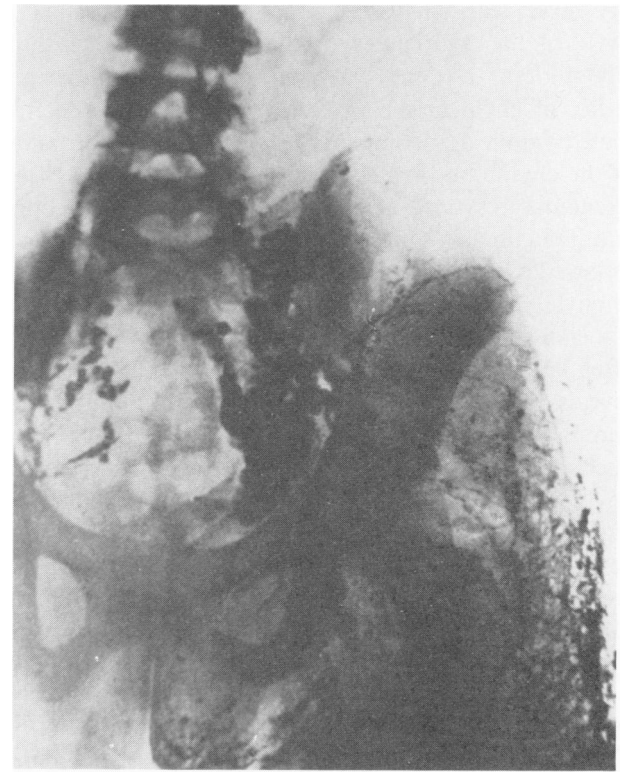

FIG. 7. Lymphography revealed extensive dilatation of the lymphatics of the whole lower limb and of the iliac region.

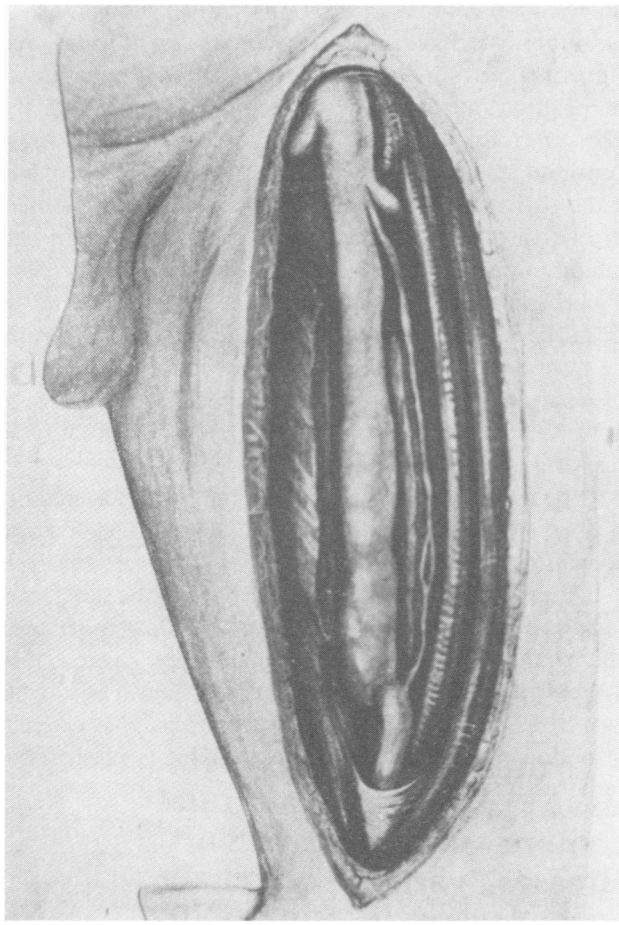

FIG. 6. Dilated retro-adductor vein.

producing chylorrhagia. Three years later, this lymphedema with reflux of chyle involved the lower limb, and we operated in 1964 and 1972. This boy suffered from Klippel and Trénaunay's syndrome, a chylothorax, chyluria, and lymphedema.

We have also treated five patients with intraosseous lymphangioma. Each of these patients also suffered from Klippel and Trénaunay's syndrome and lymphedema with reflux of chyle into the lymphatics of the legs due to a malformation of the chyliferous vessels. We have never observed an intraosseous lymphangioma in 550 patients with common lymphedema without reflux of chyle.

From 1966 to 1983, we observed 300 malformations of the chyliferous vessels and we operated upon 80. In all patients, the diagnosis was confirmed by the induced hyperlipidemia test which showed a flat total lipid curve. For the last 10 years, our hyperlipidemia test has been performed in the evaluation of Klippel and Trénaunay's syndrome, and we have noted that the malformation of the chyliferous vessels is often associated with a malformation of the deep veins. The malformation of the chyliferous vessels is responsible for a series of disorders which cannot exist independently; they are protein losing enteropathy, chyloperitoneum, chylous cysts of

the mesentery, chyluria, lymphedema with reflux of chyle in the lymphatics of the leg, chylopericardium, chylothorax, chylous cysts of the mediastinum, and reflux of chyle in the pulmonary lymphatics. Each of these complications may appear when the malformation of the chyliferous vessels is associated with Klippel and Trénaunay's syndrome. In a boy operated on for Klippel and Trénaunay's syndrome, we observed a chylothorax followed by a chyluria, and last, lymphedema with reflux of chyle. These malformations of the chyliferous vessels associated with Klippel and Trénaunay's syndrome are often latent at the time of the hyperlipidemia test which reveals them. However, it is desirable to inform the parents as to the potential complications. In 1974, we operated on a 14-year-old girl for Klippel and Trénaunay's syndrome and the hyperlipidemia test indicated a diagnosis of associated malformation of the chyliferous vessels. The parents were informed of possible secondary complications due to the abnormality of the lymphatics of the small intestine. In 1980, this patient returned for a chylothorax which required operation.

Klippel and Trénaunay's Syndrome with Gluteal Hypertrophy on the Same Side

Two of our patients suffered from Klippel and Trénaunay's syndrome associated with hypertrophy of the buttock on the same side. A young girl presented with atresia of the superficial femoral, external, and internal iliac veins on the same leg. Gluteal hypertrophy resulted from poor venous drainage.

A 6-year-old boy was operated on for popliteal Klippel and Trénaunay's syndrome, and slight edema of the buttock existed at that time. At age 30 he had a distinct hypertrophy of the buttock on the operated side. From the age of 10, he suffered from frequent and abundant gluteal chylorrhagia. The hyperlipidemia test indicated a diagnosis of malformation of the chyliferous vessels. During operation, we discovered a huge lymphatic varix (1 cm in diameter) containing chyle anterior to the iliac vein and lower vena cava. Peroperative lymphography revealed reflux of chyle in the gluteal lymphatics. Resection of these megalymphatics resulted in total disappearance of the chylorrhagia, and the patient has remained stable for 4 years with marked reduction in the size of the buttock. In this patient, gluteal hypertrophy was produced by the reflux of chyle in the lymphatics of the buttock secondary to a malformation of the lymphatics in the small intestine.

Klippel and Trénaunay's Syndrome Associated with Venous Angiomata

In September 1948, in the *Archives des maladies du coeur,* we previously described venous angiomata of the limbs. They differ anatomically, clinically, and radiologically from Klippel and Trénaunay's syndrome. The skeleton of the limb is shorter and many free intraangiomatous calculi are present.

Klippel and Trénaunay's syndrome may be associated with a localized venous angioma. There are free intraangiomatous calculi but the limb is longer and edema is evident. Venography reveals two or three localized venous angiomata with one or several malformations of the deep veins. This association is quite rare. We have seen only four such patients, three of which are reported in our book *Pathologie Veineuse* published in 1978 (pages 116–118).

Discussion

The venograms followed by corrective operation in 768 patients with Klippel and Trénaunay's syndrome have always revealed a malformation of the deep venous channels resulting in venous stasis below the obstruction. This venous stasis is responsible for the elongation of the limb and edema.

If the main vein of a limb is ligated during childhood, the resultant venous stasis produces an elongation of this limb. We have used this concept in Klippel and Trénaunay's syndrome in order to obtain elongation of the healthy limb which is often much shorter than the other limb. Klippel and Trénaunay's syndrome is more common in the lower limbs than in the arms. In half of the patients, the malformation involves the popliteal vein. An isolated abnormality of the femoral vein was found in only 16% of our patients. However, obstruction of both the popliteal and femoral veins was found in 29%. The iliac veins were implicated in only 3.3%, and the lower vena cava 0.7%. The obstruction may be due to agenesis or atresia of a segment of the main vein. However, compression by fibrous bands, abnormal muscles, or a perivenous sheath are also seen.

Klippel and Trénaunay's syndrome should preferably be operated on in childhood to avoid major edema and elongation of the limb. The posterior substitute channels of the femoral or femoroiliac malformations are sometimes associated with vesical or rectal hemorrhages which may necessitate partial cystectomy or resection of the rectosigmoid. Klippel and Trénaunay's syndrome may be associated with common lymphedema or malformation of the chyliferous vessels, but is rarely associated with venous angiomata.

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